Agency for Healthcare Research and Quality
Advancing Excellence in Health Care

NATIONAL GUIDELINE CLEARINGHOUSE

General

Guideline Title

Primary care of adults with developmental disabilities. Canadian consensus guidelines.

Bibliographic Source(s)

Sullivan WF, Berg JM, Bradley E, Cheetham T, Denton R, Heng J, Hennen B, Joyce D, Kelly M, Korossy M, Lunsky Y, McMillan S. Primary care of adults with developmental disabilities: Canadian consensus guidelines. Can Fam Physician. 2011 May;57(5):541-53, e154-68. [185 references] PubMed

Guideline Status

This is the current release of the guideline.

Regulatory Alert

FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- May 10, 2016 – Olanzapine : The U.S. Food and Drug Administration (FDA) is warning that the antipsychotic medicine olanzapine can cause a rare but serious skin reaction that can progress to affect other parts of the body. FDA is adding a new warning to the drug labels for all olanzapine-containing products that describes this severe condition known as Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS).

Recommendations

Major Recommendations

Levels of evidence (I–III) are defined at the end of the "Major Recommendations" field.

Preventive Care Checklist for Adults with Developmental Disabilities (DD)

Recommendations are listed with associated considerations under each subheading below.

General Issues in Primary Care of Adults with DD

Disparities

Consideration: Disparities in primary care exist between adults with DD and the general population. The former often have poorer health, increased morbidity, and earlier mortality. Assessments that attend to the specific health issues of adults with DD can improve their primary care.

Recommendation: Apply age- and sex-specific guidelines for preventive health care as for adults in the general population (Dubey et al., 2006; Iglar et al., 2008). Perform an annual comprehensive preventive care assessment including physical examination and use guidelines and tools adapted for adults with DD (Lennox et al., 2007). (Level I)

Etiology

Consideration: Etiology of DD is useful to establish, whenever possible, as it often informs preventive care or treatment.

Recommendation: Contact a genetics centre for referral criteria and testing protocols concerning etiologic assessment of adults whose DD is of unknown or uncertain origin (Curry et al., 1997; Diraimo et al., 2009; University of Washington, 1993). (Level III)

Consideration: Advances in genetic knowledge continue to enhance detection of etiology.

Recommendation: Consider reassessment periodically if a previous assessment was inconclusive, according to the criteria of the genetics centre (Moeschler, 2008). (Level III)

Adaptive Functioning

Consideration: Adaptive functioning can decline or improve in some adults with DD. A current assessment of intellectual and adaptive functioning helps to determine necessary care and supports, and establishes a baseline for future assessment.

Recommendation: Refer to a psychologist for assessment of functioning if the patient has never been assessed during adolescence or adulthood, or if a considerable life transition is expected (e.g., cessation of schooling or transition from middle to old age). (Level III)

Recommendation: Consider reassessment if indicated, comprehensively or in specific areas, to determine contributing factors to problem behaviour (see "Problem Behaviour," below) (The AAIDD Ad Hoc Committee on Terminology and Classification, 2010). (Level III)

Pain and Distress

Consideration: Pain and distress, often unrecognized, might present atypically in adults with DD, particularly those who have difficulty communicating. Nonspecific changes in behaviour might be the only indicator of medical illness or injury.

Recommendation: Be attentive to atypical physical cues of pain and distress using an assessment tool adapted for adults with DD (Regnard, Matthews, & Gibson, 2008; Burkitt et al., 2009). (Level III)

Consideration: Evaluation tools are available to assess the presence and intensity of pain in adults with DD.

Recommendation: Consider medical causes of changes in behaviour (e.g., urinary tract infection, dysmenorrhea, constipation, dental disease) (Regnard et al., 2007). (Level III)

Multiple or Long-term Use of Some Medications

Consideration: Multiple or long-term use of some medications by adults with DD can cause harm that is preventable.

Recommendation: Review the date of initiation, indications, dosages, and effectiveness of all medications regularly (e.g., every 3 mo) (Lennox, 2005). (Level III)

Recommendation: Determine patient adherence capacity and recommend dosettes, blister-packs, and other aids if necessary. (Level III)

Recommendation: Watch for both typical and atypical signs of adverse effects (Beange, Lennox, & Parmenter, 1999). Regularly monitor potentially toxic medications or interactions of medications (e.g., liver function tests or serum drug levels) at the recommended interval for each medication (Bhaumik & Branford, 2008). (Level III)

Recommendation: Ensure that patient and staff or caregivers are educated about appropriate use of medications, including over-the-counter, alternative, and as-needed medications. (Level III)

Abuse and Neglect

Consideration: Abuse and neglect of adults with DD occur frequently and are often perpetrated by people known to them. Behavioural indicators that might signal abuse or neglect include unexplained change in weight, noncompliance, aggression, withdrawal, depression, avoidance, poor self-esteem, inappropriate attachment or sexualized behaviour, sleep or eating disorders, and substance abuse.

Recommendation: Screen annually for risk factors (e.g., caregiver stress) and possible behavioural indicators of abuse or neglect (Fudge Schormans & Sobsey, 2007). (Level III)

Recommendation: When abuse or neglect is suspected, report to the police or other appropriate authority and address any consequent health issues (e.g., through appropriate counseling) (Fudge Schormans & Sobsey, 2007). (Level III)

Capacity for Voluntary and Informed Consent

Consideration: Capacity for voluntary and informed consent varies with the complexity and circumstances of decision making. The limited range of life experiences of some adults with DD, level of intellectual functioning, learned helplessness, and some mental health issues might impair capacity to give informed or voluntary consent. An adult with DD assessed as incapable of some aspects of decision making (e.g., understanding or judging consequences) might still be able to convey, through verbal or other means, perspectives that can inform the judgment of a substitute decision maker.

Recommendation: Always assess capacity for consent when proposing investigations or treatments for which consent is required (Friedman, 1998). (Level III)

Consideration: Communicating appropriately with adults with DD is necessary for assessing their capacity to consent and for seeking this consent.

Recommendation: Adapt the level and means of communicating to the patient's level of intellectual and adaptive functioning (van Schrojenstein Lantman-de Valk & Walsh, 2008). (Level III)

Consideration: Although some adults with DD might be incapable of giving consent, they might be able to contribute to decision making (e.g., understanding information, expressing perspectives, giving assent) with appropriate support from regular caregivers. Caregivers can also contribute to decision making. They may consent to or refuse treatment on behalf of an adult with DD who is assessed to be incapable of providing informed consent, if they are the most appropriate and available substitute decision makers according to the law.

Recommendation: Always consider the best interests of the adult with DD, including his or her perspective in pursuing or forgoing any health care intervention. Support whatever decision-making capacity is possible in adults with DD. Involve family or other caregivers to facilitate communication with, and understanding of, the adult with DD, but also be attentive to inappropriate taking over of decision making (Tuffrey-Wijne & McEnhill, 2008; Rush & Frances "Guideline 2," 2000). (Level III)

Advance Care Planning

Consideration: Advance care planning can often make a positive difference to the outcome of difficult life transitions and crises, and for end-of-life care.

Recommendation: Discuss advance care plans with adults with DD and their caregivers, especially to determine their preference of a substitute decision maker (Friedman, 1998). (Level III)

Recommendation: Record advance care plans and review them annually, or sooner in the context of a health crisis, for appropriateness to the adult with DD's present situation and for what needs to be implemented (van Schrojenstein Lantman-de Valk & Walsh, 2008). (Level III)

Interdisciplinary Health Care

Consideration: Interdisciplinary health care is effective in addressing the complex needs of adults with DD. Ideally this would involve a family physician, nurse, and other health practitioners as required, with a coordinator, who might be the family physician, to ensure continuity of care.

Recommendation: Involve other available health professionals as needed (Crocker, 2006). To address complex physical, behavioural, or mental health needs, consult available regional service coordination agencies or specialized interdisciplinary teams (Ministry of Community and Social Services, 2008; Ministry of Community and Social Services, 2009). (Level III)

Physical Health Guidelines for Adults with DD

Physical Inactivity and Obesity

Consideration: Physical inactivity and obesity are prevalent among adults with DD and are associated with adverse outcomes, including

cardiovascular disease, diabetes, osteoporosis, constipation, and early mortality. Being underweight, with its attendant health risks, is also common.

Recommendation: Monitor weight and height regularly and assess risk status using body mass index, waist circumference, or waist-hip ratio measurements (Health Canada, 2003; Bhaumik et al., 2008). (Level II)

Consideration: A health promotion program can improve attitudes toward physical activity and satisfaction with life.

Recommendation: Counsel patients and their caregivers annually or more frequently, if indicated, regarding guidelines for nutrition and physical fitness and how to incorporate regular physical activity into daily routines. Refer to dietitian if indicated (Marks, Sisirak, & Heller, 2010; Healthy Living Unit, Public Health Agency of Canada, 2003; Hamilton et al., 2007; Henderson et al., 2008). (Level II)

Vision and Hearing Impairments

Consideration: Vision and hearing impairments among adults with DD are often underdiagnosed and can result in substantial changes in behaviour and adaptive functioning.

Recommendation: Perform office-based screening of vision and hearing (e.g., Snellen eye chart, whispered voice test) annually as recommended for average-risk adults, and when symptoms or signs of visual or hearing problems are noted, including changes in behaviour and adaptive functioning (Beange, Lennox, & Parmenter, 1999; Evenhuis & Natzgam, 1997). (Level III)

Recommendation: Refer for vision assessment to detect glaucoma and cataracts every 5 y after age 45 (Evenhuis & Natzgam, 1997). (Level III)

Recommendation: Refer for hearing assessment if indicated by screening and for age-related hearing loss every 5 y after age 45 (Evenhuis & Natzgam, 1997). (Level III)

Recommendation: Screen for and treat cerumen impaction every 6 mo (Crandell & Roeser, 1993; Roland et al., 2008). (Level III)

Dental Disease

Consideration: Dental disease is among the most common health problems in adults with DD owing to their difficulties in maintaining oral hygiene routines and accessing dental care. Changes in behaviour can be the result of discomfort from dental disease.

Recommendation: Promote regular oral hygiene practices and other preventive care (e.g., fluoride application) by a dental professional (Ismail, Lewis, & Dingle, 1994; Lewis & Ismail, 1994; Dougherty & MacRae, 2006; Glassman & Miller, 2003). (Level I)

Cardiac Disorders

Consideration: Cardiac disorders are prevalent among adults with DD. Risk factors for coronary artery disease include physical inactivity, obesity, smoking, and prolonged use of some psychotropic medications.

Recommendation: When any risk factor is present, screen for cardiovascular disease earlier and more regularly than in the general population and promote prevention (e.g., increasing physical activity, reducing smoking) (Wallace & Schluter, 2008). (Level III)

Consideration: Some adults with DD have congenital heart disease and are susceptible to bacterial endocarditis.

Recommendation: Refer to a cardiologist or adult congenital heart disease clinic (Canadian Adult Congenital Health Network, 2009). (Level III)

Recommendation: Follow guidelines for antibiotic prophylaxis for those few patients who meet revised criteria (Wilson et al., 2007). (Level II)

Respiratory Disorders

Consideration: Respiratory disorders (e.g., aspiration pneumonia) are among the most common causes of death for adults with DD. Swallowing difficulties are prevalent in those patients with neuromuscular dysfunction or taking certain medications with anticholinergic side effects, and they might result in aspiration or asphyxiation.

Recommendation: Screen at least annually for possible signs of swallowing difficulty and overt or silent aspiration (e.g., throat clearing after swallowing, coughing, choking, drooling, long mealtimes, aversion to food, weight loss, frequent chest infections). Refer as appropriate (Chadwick & Jolliffe, 2009). (Level III)

Gastrointestinal and Feeding Problems

Consideration: Gastrointestinal and feeding problems are common among adults with DD. Presenting manifestations are often different than in the

general population and might include changes in behaviour or weight.

Recommendation: Screen annually for manifestations of gastroesophageal reflux disease (GERD) and manage accordingly. If introducing medications that can aggravate GERD, monitor more frequently for related symptoms (Wallace et al., 2004; Böhmer et al., 2000). (Level III)

Recommendation: If there are unexplained gastrointestinal findings or changes in behaviour or weight, investigate for constipation, GERD, peptic ulcer disease, and pica (Morad et al., 2007; Böhmer et al., 2000). (Level II)

Consideration: Adults with DD might have an increased risk of *Helicobacter pylori* infection related to factors such as having lived in a group home, rumination, or exposure to saliva or feces due to personal behaviour or environmental contamination.

Recommendation: Screen for *H pylori* infection in symptomatic adults with DD or asymptomatic ones who have lived in institutions or group homes. Consider retesting at regular intervals (e.g., 3–5 y) (Wallace et al., 2004). (Level III)

Recommendation: Consider urea breath testing, fecal antigen testing, or serologic testing depending on the indication, availability, and tolerability of the test (Wallace et al., 2004; Bourke et al., 2005). (Level III)

Sexuality

Consideration: Sexuality is an important issue that is often not considered in the primary care of adolescents and adults with DD.

Recommendation: Discuss the patient's or caregiver's concerns about sexuality (e.g., menstruation, masturbation, fertility and genetic risks, contraception, menopause) and screen for potentially harmful sexual practices or exploitation. Offer education and counseling services adapted for those with DD (Wilkinson & Cerreto, 2008; Cox, Signore, & Quint, 2011). (Level III)

Musculoskeletal Disorders

Consideration: Musculoskeletal disorders (e.g., scoliosis, contractures, and spasticity, which are possible sources of unrecognized pain) occur frequently among adults with DD and result in reduced mobility and activity, with associated adverse health outcomes.

Recommendation: Promote mobility and regular physical activity (Marks, Sisirak, & Heller, 2010; O'Neil et al., 2006). (Level III)

Recommendation: Consult a physical or occupational therapist regarding adaptations (e.g., wheelchair, modified seating, splints, orthotic devices) and safety (O'Neil et al., 2006). (Level III)

Consideration: Osteoporosis and osteoporotic fractures are more prevalent and tend to occur earlier in adults with DD than in the general population. In addition to aging and menopause, risk factors include severity of DD, low body weight, reduced mobility, increased risk of falls, smoking, hypogonadism, hyperprolactinemia, the presence of particular genetic syndromes (e.g., Down and Prader-Willi), and long-term use of certain drugs (e.g., glucocorticoids, anticonvulsants, injectable long-acting progesterone in women). Diagnosis and management of osteoporosis related to the side effects of current treatments can be challenging in adults with DD.

Recommendation: Periodically assess risk of developing osteoporosis in all age groups of male and female patients with DD. Those at high risk warrant regular screening starting in early adulthood (Brown & Josse, 2002; Zylstra et al., 2008). (Level III)

Recommendation: Recommend early and adequate intake or supplementation of calcium and vitamin D unless contraindicated (e.g., in Williams syndrome) (Brown & Josse, 2002). (Level III)

Consideration: Osteoarthritis is becoming more common with increasing life expectancy and weight gain, posing diagnostic and treatment difficulties.

Recommendation: Be aware of osteoarthritis as a possible source of pain (Haveman et al., 2010). (Level III)

Epilepsy

Consideration: Epilepsy is prevalent among adults with DD and increases with the severity of the DD. It is often difficult to recognize, evaluate, and control, and has a pervasive effect on the lives of affected adults and their caregivers.

Recommendation: Refer to guidelines for management of epilepsy in adults with DD (Kerr et al., 2009). (Level III)

Recommendation: Review seizure medication regularly (e.g., every 3–6 mo). Consider specialist consultation regarding alternative medications when seizures persist, and possible discontinuation of medications for patients who become seizure-free (Kerr et al., 2009). (Level III)

Recommendation: Educate patients and caregivers about acute management of seizures and safety-related issues (Epilepsy Canada, 2009). (Level III)

Endocrine Disorders

Consideration: Endocrine disorders (e.g., thyroid disease, diabetes, and low testosterone) can be challenging to diagnose in adults with DD. Adults with DD have a higher incidence of thyroid disease compared with the general population.

Recommendation: Monitor thyroid function regularly. Consider testing for thyroid disease in patients with symptoms (including changes in behaviour and adaptive functioning) and at regular intervals (e.g., 1–5 y) in patients with elevated risk of thyroid disease (e.g., Down syndrome) (Beange, Lennox, & Parmenter, 1999). (Level III)

Recommendation: Establish a thyroid baseline and test annually for patients taking lithium or atypical or second-generation antipsychotic drugs (Bhaumik & Branford, 2008). (Level III)

Consideration: Currently there is no clear evidence of increased prevalence of diabetes in adults with DD, with some exceptions (e.g., Down syndrome). Diabetes management guidance has been developed for adults with DD and their care providers.

Recommendation: Consider screening for diabetes in adults with DD who are obese or who have sedentary lifestyles or hyperlipidemia. (Level III)

Consideration: Limited available data suggest that hypogonadism is common among men with DD. Substantial data are available on hypogonadism associated with specific syndromes (e.g., Prader-Willi syndrome).

Recommendation: Consider screening for hypogonadism and testosterone level at least once after full puberty is achieved, ideally at around age 18 y, and refer as appropriate if low levels are found (McElduff & Beange, 2003; McElduff, Center, & Beange, 2003). (Level III)

Infectious Disease Prevention and Screening

Consideration: Infectious disease prevention and screening. Even though immunization is a crucial component of preventive care, adults with DD might have limited awareness of immunizations.

Recommendation: Follow guidelines for routine immunization of adults (Langley & Faughnan, 2004; National Advisory Committee on Immunization [NACI], 2006). (Level III)

Recommendation: Ensure influenza and *Streptococcus pneumoniae* vaccinations are current and offered when appropriate ("23-valent pneumococcal polysaccharide vaccine," 2008). (Level III)

Recommendation: Discuss the human papillomavirus vaccine with female patients with DD between the ages of 9 and 26 y and, if appropriate, their substitute decision makers (NACI, 2007). (Level III)

Consideration: It is important to screen for infectious diseases (e.g., hepatitis B, human immunodeficiency virus [HIV], and *H pylori*) in adults with DD.

Recommendation: Screen for infectious diseases based on the patient's risk factors for exposure (for recommendations on *H pylori* see "Gastrointestinal and Feeding Problems," above). (Level III)

Consideration: Some adults with DD have an increased risk of exposure to infectious diseases (e.g., hepatitis A and B).

Recommendation: Offer hepatitis A and B screening and immunization to all at-risk adults with DD, (NACI, 2007; Mast et al., 2006; Advisory Committee on Immunization Practices, 2006) including those who take potentially hepatotoxic medications or who have ever lived in institutions or group homes (NACI, 2006). (Level III)

Cancer Screening

Consideration: Cancer screening is an essential aspect of preventive care. However, adults with DD are less likely than those in the general population to be included in preventive screening programs such as cervical screening, breast examination, mammography, and digital rectal examination. They are also less likely to do self-examination or to report abnormalities. Colorectal cancer risk is considerably greater for women than for men with DD.

Recommendation: Perform regular cervical screening for all women who have been sexually active (Quint & Elkins, 1997). (Level I)

Recommendation: Perform annual breast screening, including mammography, for female patients with DD aged 50–69 y (Morrison, 1994). (Level

III)

Recommendation: Perform an annual testicular examination for all male patients with DD (Elford, 1994). (Level III)

Recommendation: Screen for prostate cancer annually using digital rectal examination from age 45 y for all male patients with DD (Feightner, 1994). (Level II)

Recommendation: Screen for colon cancer regularly in all adult patients with DD older than 50 y (Sullivan et al., 2004; Leddin et al., 2004). (Level I)

Behavioural and Mental Health Guidelines for Adults with DD

Problem Behaviour

Consideration: Problem behaviour, such as aggression and self-injury, is not a psychiatric disorder but might be a symptom of a health-related disorder or other circumstance (e.g., insufficient supports).

Recommendation: Before considering a psychiatric diagnosis, assess and address sequentially possible causes of problem behaviour, including physical (e.g., infections, constipation, pain), environmental (e.g., changed residence, reduced supports), and emotional factors (e.g., stress, trauma, grief) (Bradley & Hollins, 2010). (Level II)

Consideration: Problem behaviours sometimes occur because environments do not meet the developmental needs of the adult with DD.

Recommendation: Facilitate "enabling environments" to meet these unique developmental needs as they will likely diminish or eliminate these problem behaviours (Banks et al., 2007). (Level III)

Consideration: Despite the absence of an evidence base, psychotropic medications are regularly used to manage problem behaviours among adults with DD. Antipsychotic drugs should no longer be regarded as an acceptable routine treatment of problem behaviours in adults with DD.

Recommendation: Regularly audit the use of prescribed psychotropic medication, including those used as needed (Deb, Clarke, & Unwin, 2006). Plan for a functional analysis (typically performed by a behavioural therapist or psychologist) and interdisciplinary understanding of problem behaviours. Review with care providers psychological, behavioural, and other nonmedication interventions to manage problem behaviours. Consider reducing and stopping, at least on a trial basis, medications not prescribed for a specific psychiatric diagnosis (Deb et al., 2009). (Level III)

Psychiatric Disorders

Consideration: Psychiatric disorders and emotional disturbances are substantially more common among adults with DD, but their manifestations might mistakenly be regarded as typical for people with DD (i.e., "diagnostic overshadowing"). Consequently, coexisting mental health disturbances might not be recognized or addressed appropriately.

Recommendation: When screening for psychiatric disorder or emotional disturbance, use tools developed for adults with DD according to their functioning level (e.g., Aberrant Behaviour Checklist-Community [ABC-C]; Psychiatric Assessment Schedule for Adults with DD [PAS-ADD]) (Aman, Burrow, & Wolford, 1995; Moss et al., 1998; Mohr & Costello, 2007; Perez-Achiaga, Nelson, & Hassiotis, 2009). (Level III)

Consideration: Increased risk of particular developmental, neurologic, or behavioural manifestations and emotional disturbances (i.e., "behavioural phenotypes") is associated with some DD syndromes.

Recommendation: Consult available information on behavioural phenotypes in adults with DD due to specific syndromes (O'Brien, 2002; Society for the Study of Behavioural Phenotypes, 2009). (Level III)

Consideration: Establishing a diagnosis of a psychiatric disorder in adults with DD is often complex and difficult, as these disorders might be masked by atypical symptoms and signs. In general, mood, anxiety, and adjustment disorders are underdiagnosed and psychotic disorders are overdiagnosed in adults with DD.

Recommendation: When psychiatric disorder is suspected, seek interdisciplinary consultation from clinicians knowledgeable and experienced in DD. (Level III)

Psychotic Disorders

Consideration: Psychotic disorders are very difficult to diagnose when delusions and hallucinations cannot be expressed verbally. Developmentally appropriate fantasies and imaginary friends might be mistaken for delusional ideation, and self-conversation for hallucination.

Recommendation: Seek interdisciplinary input from specialists in psychiatry, psychology, and speech-language pathology with expertise in DD to help clarify diagnoses in patients with limited or unusual use of language (Bradley et al., 2009; Craft, Bicknell, & Hollins, 1985; Summers et al., "The interdisciplinary," 2002). (Level III)

Input and Assistance from Adults with DD and Their Caregivers

Consideration: Input and assistance from adults with DD and their caregivers are vital for a shared understanding of the basis of problem behaviours, emotional disturbances, and psychiatric disorders, and for effectively developing and implementing treatment and interventions.

Recommendation: Establish a shared way of working with patients and caregivers. Seek input, agreement, and assistance in identifying target symptoms and behaviours that can be monitored. (Level III)

Recommendation: Use tools (e.g., sleep charts, antecedent–behaviour-consequence [ABC] charts) to aid in assessing and monitoring behaviour and intervention outcomes (Summers et al. "Comprehensive screening," 2002; Hurley, 1997). (Level III)

Interventions Other Than Medication

Consideration: Interventions other than medication are usually effective for preventing or alleviating problem behaviours.

Recommendation: To reduce stress and anxiety that can underlie some problem behaviours, emotional disturbances, and psychiatric disorders, consider such interventions as addressing sensory issues (e.g., underarousal, overarousal, hypersensitivity), environmental modification, education and skill development, communication aids, psychological and behaviour therapies, and caregiver support (Bradley et al., 2009). (Level III)

Recommendation: Cognitive behavioural therapy can be effective in decreasing anger and treating anxiety and depression in adults with DD (McCabe, McGillivray, & Newton, 2006; Taylor, Lindsay, & Willner, 2008). (Level III)

Recommendation: There is increasing evidence of the efficacy of psychotherapy for emotional problems (e.g., related to grief, abuse, trauma) that might underlie aggression, anxiety, and other such states (Hollins & Sinason, 2000; Hubert & Hollins, 2006; McGinnity et al., 2004; Sequeira, Howlin, & Hollins, 2003; Cottis, 2008). (Level III)

Psychotropic Medications

Consideration: Psychotropic medications (e.g., antidepressants) are effective for robust diagnoses of psychiatric disorders in adults with DD as in the general population.

Recommendation: When psychiatric diagnosis is confirmed after comprehensive assessment, consider psychotropic medication along with other appropriate interventions as outlined in "Intervention Other Than Medication," above (Kalachnik et al., 1998). (Level III)

Consideration: Psychotropic medications, however, can be problematic for adults with DD and should therefore be used judiciously. Patients might be taking multiple medications and can thus be at increased risk of adverse medication interactions. Some adults with DD might have atypical responses or side effects at low doses. Some cannot describe harmful or distressing effects of the medications that they are taking.

Recommendation: "Start low, go slow" in initiating, increasing, or decreasing doses of medications (Rush & Frances "Guideline 4," 2000). (Level III)

Recommendation: Arrange to receive regular reports from patients and their caregivers during medication trials in order to monitor safety, side effects, and effectiveness (Deb et al., 2009). (Level III)

Recommendation: In addition to reviews every 3 mo (see "Multiple or Long-term Use of Some Medications," above), also review the psychiatric diagnosis and the appropriateness of prescribed medications for this diagnosis whenever there is a behavioural change (Bhaumik & Branford, 2008; Deb et al., 2009). (Level III)

Consideration: When unable to pinpoint a specific psychiatric diagnosis, behaviours of concern might serve as index behaviours against which to conduct a trial of medications.

Recommendation. Having excluded physical, emotional, and environmental contributors to the behaviours of concern, a trial of medication appropriate to the patient's symptoms might be considered. (Level III)

Antipsychotic Medications

Consideration: Antipsychotic medications are often inappropriately prescribed for adults with behaviour problems and DD. In the absence of a robust diagnosis of psychotic illness, antipsychotic medications should not be regarded as routine treatments of problem behaviours in adults with

DD.

Recommendation: Do not use antipsychotic medication as a first-line treatment of problem behaviours without a confirmed robust diagnosis of schizophrenia or other psychotic disorder (Tyrer et al., 2008). (Level III)

Consideration: Antipsychotic medications increase risk of metabolic syndrome and can have other serious side effects (e.g., akathisia, cardiac conduction problems, swallowing difficulties, bowel dysfunction).

Recommendation: Carefully monitor for side effects of antipsychotic medication, including metabolic syndrome. Educate patients and caregivers to incorporate a healthy diet and regular exercise into their lifestyle (Bhaumik & Branford, 2008). (Level III)

Recommendation: Reassess the need for ongoing antipsychotic medications at regular intervals and consider dose reduction or discontinuation when appropriate (also see "Multiple or Long-term Use of Some Medications" and "Psychotropic Medications," above) (Bhaumik & Branford, 2008). (Level III)

Behavioural Crises

Consideration: Behavioural crises can occasionally arise that might need management in an emergency department.

Recommendation: When psychotropic medications are used to ensure safety during a behavioural crisis, ideally such use should be temporary (no longer than 72 h). (Level III)

Recommendation: Debrief with care providers in order to minimize the likelihood of recurrence. This should include a review of crisis events and responses (e.g., medication, de-escalation measures), and identification of the possible triggers and underlying causes of the behavioural crisis (Deb et al., 2009; Bradley & Lofchy, 2005). (Level III)

Recommendation: If the patient is at risk of recurrent behavioural crises, involve key stakeholders, including local emergency department staff, to develop a proactive, integrated emergency response plan (Bradley & Lofchy, 2005). (Level III)

Alcohol or Drug Abuse

Consideration: Alcohol or drug abuse is less common among adults with DD than in the general population, but the former might have more difficulty moderating their intake and experience more barriers to specialized rehabilitation services.

Recommendation: Screen for alcohol and drug abuse as part of the annual health examination. (Level III)

Dementia

Consideration: Dementia is important to diagnose early, especially in adults with Down syndrome who are at increased risk. Diagnosis might be missed because changes in emotion, social behaviour, or motivation can be gradual and subtle. A baseline of functioning against which to measure changes is needed.

Recommendation: For patients at risk of dementia, assess or refer for psychological testing to establish a baseline of cognitive, adaptive, and communicative functioning. Monitor with appropriate tools (Royal College of Psychiatrists & British Psychological Society, 2009). (Level III)

Consideration: Differentiating dementia from depression and delirium can be especially challenging.

Recommendation: Educate family and other care providers about early signs of dementia. When signs are present, investigate for potential reversible causes of dementia. (Level III)

Recommendation: Consider referral to the appropriate specialist (i.e., psychiatrist, neurologist) if it is unclear whether symptoms and behaviour are due to emotional disturbance, psychiatric disorder, or dementia (Royal College of Psychiatrists & British Psychological Society, 2009). (Level III)

Definitions:

Criteria for Assigning Levels of Evidence

Level I: At least 1 properly conducted randomized controlled trial, systematic review, or meta-analysis

Level II: Other comparison trials, non-randomized, cohort, case control, or epidemiologic studies, and preferably more than 1 study

Level III: Expert opinion or consensus statements

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Developmental disabilities

Guideline Category

Counseling

Diagnosis

Evaluation

Management

Prevention

Risk Assessment

Screening

Treatment

Clinical Specialty

Family Practice

Preventive Medicine

Intended Users

Advanced Practice Nurses

Physician Assistants

Physicians

Guideline Objective(s)

- To update the 2006 Canadian guidelines for primary care of adults with developmental disabilities (DD)
- To make practical recommendations based on current knowledge to address the particular health issues of adults with DD

Target Population

Adults with developmental disabilities

Interventions and Practices Considered

Prevention

1. Application of age- and sex-specific prevention guidelines
2. Annual comprehensive preventive care assessment
3. Antibiotic prophylaxis for those at risk of bacterial endocarditis
4. Immunizations for infectious disease
5. Vitamin D and calcium supplementation

Screening

1. Screening for cardiovascular disease
2. Screen for respiratory problems, including swallowing difficulty and overt or silent aspiration
3. Screen annually for manifestations of gastrointestinal and feeding problems: gastroesophageal reflux disease (GERD) and *Helicobacter pylori* infection
4. Screening for risk factors that might signal abuse or neglect
5. Office-based screening of vision and hearing
6. Screen for potentially harmful sexual practices or exploitation
7. Screening for endocrine disorders: diabetes, hypogonadism and testosterone level
8. Screen for infectious diseases: hepatitis A and B
9. Cancer screening: cervical, breast, testicular, and prostate cancers
10. Screening for psychiatric disorder or emotional disturbance
    - Aberrant Behaviour Checklist-Community (ABC-C)
    - Psychiatric Assessment Schedule for Adults with Developmental Disabilities (PAS-ADD)
11. Screen for alcohol and drug abuse

Evaluation

1. Referral to psychologist for assessment of intellectual and adaptive functioning
2. Use of an evaluation tool to assess the presence and intensity of pain
3. Consideration of medical causes of behaviour changes
4. Assessment of capacity for consent
5. Monitoring anthropometric measurements
6. Assessment for osteoporosis risk
7. Assessment of problem behaviour
8. Use of tools to assess and monitor behaviour and intervention outcomes
    - Sleep charts
    - Antecedent–behaviour-consequence (ABC) charts
9. Assessment or referral for psychological testing patients at risk of dementia

Management/Treatment

1. Regular review of all medications, including dosing, effectiveness, patient adherence, and the possibility of stopping or reducing medications
2. Adaptation of level and means of communication to patient level of understanding
3. Advance care planning
4. Interdisciplinary health care
5. Counseling regarding nutrition and physical activity
6. Referral to specialists as necessary
7. Adaptation aids, including wheelchairs, modified seating, splints, and enabling environments
8. Management of epilepsy, including medication review and education of patients and families about seizures
9. Monitor thyroid function regularly, with establishment of a baseline
10. Cognitive behavioural therapy and psychotherapy
11. Psychotropic medications
12. Antipsychotic medications with careful monitoring
13. Involvement of patient and caregivers in management
14. Education of family and caregiver about early signs of dementia

Major Outcomes Considered

- Quality of primary care of adults with developmental disabilities
- Effectiveness of screening/assessment tools
- Effectiveness of diagnostic strategies
- Effectiveness of treatment strategies
- Level of function
- Health status
- Safety, side effects, and effectiveness of medications
- Access to health care
- Quality of life

Methodology

Methods Used to Collect/Select the Evidence

Hand-searches of Published Literature (Primary Sources)

Hand-searches of Published Literature (Secondary Sources)

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

A librarian familiar with research on developmental disabilities undertook ongoing, comprehensive electronic searches in PubMed and PsycINFO for publications in English from 1990 to 2010 that were indexed under, or contained in their title, abstract, or text, the terms *mental retardation, intellectual disability (disabilities)*, or *developmental disability (disabilities)*. Publications from Great Britain were also searched for the terms *learning difficulties, learning disability (disabilities)*, or *learning disorders*. These were cross-referenced with a long list of physical and mental health key words relating to medical assessment, diagnosis, treatment, prognosis, health care access, need, planning, services, and delivery. The search was then expanded to include specific health issues highlighted in the 2006 Guidelines. In addition, the librarian undertook manual searches using cited references in Scopus and Internet searches for relevant publications that had not been indexed by any of the above-mentioned electronic databases. Search results were downloaded to and organized in an electronic database management system known as RefWorks.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Criteria for Assigning Levels of Evidence

Level I: At least 1 properly conducted randomized controlled trial, systematic review, or meta-analysis

Level II: Other comparison trials, non-randomized, cohort, case control, or epidemiologic studies, and preferably more than 1 study

Level III: Expert opinion or consensus statements

Methods Used to Analyze the Evidence

Systematic Review

Description of the Methods Used to Analyze the Evidence

The working group reviewed published supporting evidence for all the guidelines, including those from supplementary electronic and manual searches for publications undertaken after the colloquium to address particular issues that were not foreseen in the original literature searches. The working group judged the level of evidence supplied for any modified or new guidelines, using the classification scheme adopted in the 2006 Guidelines (see the "Rating Scheme for the Strength of Evidence" field).

Most of the recommendations specifically concerning adults with developmental disabilities in the updated guidelines are supported by Level III evidence based on expert opinion or published consensus statements. Three are based on randomized controlled trials, systematic reviews, or meta-analysis (Level I evidence), and 7 are based on less methodologically rigorous studies (Level II). Even when Level I or II evidence for recommendations for the general population was found, but no Level I or II evidence relating specifically to people with developmental disabilities, it was thought prudent, in view of differences between these 2 groups, to reject, adapt, or formulate new guidelines based on expert opinion (Level III evidence) for these guidelines.

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

A consensus development method was used to update the 2006 Guidelines. This consisted of 2 steps: meticulous electronic and manual searches for relevant publications and discussion of recommended changes to the 2006 Guidelines by knowledgeable and experienced Canadian clinicians and researchers on primary health care of adults with developmental disabilities who participated in a consensus colloquium in March 2009 and in a subsequent working group.

Two family physicians, a psychologist, and a psychiatrist drew on the database of search results and on comments regarding the 2006 Guidelines gained from various users and reviewers. They were each assigned a section of the 2006 Guidelines for which they were to propose revisions.

Participants who helped to formulate the 2006 Guidelines and others who had completed training courses on the guidelines between 2006 and 2009 were invited to a day-long colloquium in Toronto, Ontario, in March 2009. Among the 39 participants were practitioners in family medicine, nursing, pediatrics, psychiatry, psychology, occupational therapy, and speech-language pathology. Before the colloquium, all had access to the librarian's entries into the RefWorks database and received a summary of feedback from users and reviewers of the 2006 Guidelines. The prepared proposals for revisions were discussed in small groups and in plenary sessions, and a summary of accepted revisions was presented and discussed at the end of the colloquium in relation to the priority criteria adopted in the 2006 Guidelines (see the table below). A working group consisting of 7 participants, with a family physician in the leading role, met monthly between March 2009 and March 2010 to draft the first version of the updated guidelines. They incorporated into the 2006 Guidelines the changes discussed and accepted during the colloquium.

Guideline Priority Criteria

Criteria	Explanation
Importance	Guidelines that address the most prevalent health issues for people with developmental disabilities, especially the leading causes of ill health and death
Disparity	Guidelines that address an issue that would not be identified by public health initiatives or illness prevention measures that target the general population
Usefulness	Guidelines that can be practically implemented and evaluated; these refer to health problems that are easy to detect, for which the means of prevention and care are readily available, and which have health outcomes that can be monitored
Information	Guidelines that are supported by reliable clinical information and research evidence

Adapted from POMONA Partnership. POMONA II. Health indicators for people with intellectual disability: using an indicator set (final report). POMONA Partnership; 2008. Available from: www.pomonaproject.org.

Rating Scheme for the Strength of the Recommendations

Not applicable

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation

The first draft of the updated guidelines was circulated for review by participants in the colloquium as well as several invited consultants who were unable to attend the colloquium. Based on the feedback received, the working group prepared the second and final draft between March and October of 2010. The final draft was sent to participants in the colloquium and review process for their approval; it was then submitted for review for publication.

Evidence Supporting the Recommendations

References Supporting the Recommendations

23-valent pneumococcal polysaccharide vaccine. WHO position paper. Wkly Epidemiol Rec. 2008 Oct 17;83(42):373-84. PubMed

Advisory Committee on Immunization Practices (ACIP), Fiore AE, Wasley A, Bell BP. Prevention of hepatitis A through active or passive immunization: recommendations of the Advisory Committee on Immunization Practices (ACIP). MMWR Recomm Rep. 2006 May 19;55(RR-7):1-23. [197 references] PubMed

Aman MG, Burrow WH, Wolford PL. The Aberrant Behavior Checklist-Community: factor validity and effect of subject variables for adults in group homes. Am J Ment Retard. 1995 Nov;100(3):283-92. PubMed

Banks R, Bush A, Baker P, Bradshaw J, Carpenter P, Deb S, et al. Challenging behaviour: a unified approach. London (UK): Royal College of Psychiatrists, British Psychological Society, Royal College of Speech and Language Therapists; 2007.

Beange H, Lennox N, Parmenter TR. Health targets for people with an intellectual disability. J Intellect Dev Disabil. 1999;24(4):283-97.

Bhaumik S, Branford D. Prescribing practice and physical monitoring. In: The Frith prescribing guidelines for adults with intellectual disability. 2nd ed. London (UK): HealthComm UK Ltd; 2008. p. 7-32.

Bhaumik S, Watson JM, Thorp CF, Tyrer F, McGrother CW. Body mass index in adults with intellectual disability: distribution, associations and service implications: a population-based prevalence study. J Intellect Disabil Res. 2008 Apr;52(Pt 4):287-98. PubMed

Bohmer CJ, Klinkenberg-Knol EC, Niezen-de Boer MC, Meuwissen SG. Gastroesophageal reflux disease in intellectually disabled individuals: how often, how serious, how manageable. Am J Gastroenterol. 2000 Aug;95(8):1868-72. [67 references] PubMed

Bourke B, Ceponis P, Chiba N, Czinn S, Ferraro R, Fischbach L, Gold B, Hyunh H, Jacobson K, Jones NL, Koletzko S, Lebel S, Moayyedi P, Ridell R, Sherman P, van Zanten S, Beck I, Best L, Boland M, Bursey F, Chaun H, Cooper G, Craig B, Creuzenet C, Critch J, Govender K, Hassall E, Kaplan A, Keelan M, Noad G, Robertson M, Smith L, Stein M, Taylor D, Walters T, Persaud R, Whitaker S, Woodland R, Canadian Helicobacter Study Group. Canadian Helicobacter Study Group Consensus Conference: Update on the approach to Helicobacter pylori infection in children and adolescents--an evidence-based evaluation. Can J Gastroenterol. 2005 Jul;19(7):399-408. [93 references] PubMed

Bradley E, Lofchy J. Learning disability in the accident and emergency department. Adv Psychiatr Treat. 2005;11:45-57.

Bradley EA, Goody R, McMillan S, Levitas A. Common mental disorders (depression, anxiety, OCD, PTSD). In: Hassiotis A, Barron DA, Hall I, editor(s). Intellectual disability psychiatry: a practical handbook. Chichester, England: Wiley-Blackwell; 2009. p. 51-66.

Bradley EA, Hollins S. Assessment of patients with intellectual disabilities. In: Goldbloom DS, editor(s). Psychiatric clinical skills. 1st ed. Toronto (ON): Centre for Addiction and Mental Health; 2010. p. 257-76.

Brown JP, Josse RG. 2002 clinical practice guidelines for the diagnosis and management of osteoporosis in Canada. CMAJ. 2002 Nov 12;167(10 Suppl):S1-S34. PubMed

Burkitt C, Breau LM, Salsman S, Sarsfield Turner T, Mullan R. Pilot study of the feasibility of the non-communicating children's pain checklist revised for pain assessment in adults with intellectual disabilities. J Pain Manage. 2009;2(1):37-49.

Canadian Adult Congenital Health Network. Find a centre. [internet]. Mississauga (ON): CACH Network; 2009 [accessed 2009 Sep 24].

Chadwick DD, Jolliffe J. A descriptive investigation of dysphagia in adults with intellectual disabilities. J Intellect Disabil Res. 2009 Jan;53(1):29-43. PubMed

Cottis T. Intellectual disability, trauma, and psychotherapy. New York (NY): Routledge; 2008.

Cox RL, Signore C, Quint E. Interactive site for clinicians serving women with disabilities. [internet]. Washington (DC): American Congress of Obstetricians and Gynecologists; 2011 [accessed 2010 May 11].

Craft MJ, Bicknell DJ, Hollins S. Mental handicap: a multi-disciplinary approach. London (UK): Bailliere Tindall; 1985.

Crandell CC, Roeser RJ. Incidence of excessive/impacted cerumen in individuals with mental retardation: a longitudinal investigation. Am J Ment Retard. 1993 Mar;97(5):568-74. PubMed

Crocker AC. Systems of medical care delivery. In: Rubin IL, Crocker AC, editor(s). Medical care for children and adults with developmental

disabilities. 2nd ed. Baltimore (MD): Paul H. Brookes Publishing Co.; 2006. p. 57-9.

Curry CJ, Stevenson RE, Aughton D, Byrne J, Carey JC, Cassidy S, Cunniff C, Graham JM Jr, Jones MC, Kaback MM, Moeschler J, Schaefer GB, Schwartz S, Tarleton J, Opitz J. Evaluation of mental retardation: recommendations of a Consensus Conference: American College of Medical Genetics. Am J Med Genet. 1997 Nov 12;72(4):468-77. [70 references] PubMed

Deb S, Clarke D, Unwin G. Using medication to manage behaviour problems among adults with a learning disability: quick refernce guide (QRG). Birmingham (UK): University of Birmingham; 2006.

Deb S, Kwok H, Bertelli M, Salvador-Carulla L, Bradley E, Torr J, Barnhill J, Guideline Development Group of the WPA Section on Psychiatry of Intellectual. International guide to prescribing psychotropic medication for the management of problem behaviours in adults with intellectual disabilities. World Psychiatry. 2009 Oct;8(3):181-6. PubMed

Diraimo J, Provincial IODE Genetics Resource Centre, London Health Scienes Centre. Genetic resources Ontario. [internet]. London (ON): London Health Sciences Centre; 2009 [accessed 2009 Apr 28].

Dougherty N, MacRae R. Providing dental care to patients with developmental disabilities. An introduction for the private practitioner. N Y State Dent J. 2006 Mar-Apr;72(2):29-32. PubMed

Dubey V, Mathew R, Iglar K, Moineddin R, Glazier R. Improving preventive service delivery at adult complete health check-ups: the Preventive health Evidence-based Recommendation Form (PERFORM) cluster randomized controlled trial. BMC Fam Pract. 2006;7:44. PubMed

Elford RW. Screening for testicular cancer. In: Canadian Task Force on the Periodic Health Examination. Canadian guide to clinical preventive health care. Ottawa (ON): Health Canada; 1994. p. 892-8.

Epilepsy Canada. Coping. [internet]. Toronto (ON): Epilepsy Canada; [accessed 2009 Sep 29].

Evenhuis HM, Natzgam LM. IASSID international consensus statement: early identification of hearing and visual impairment in children and adults with an intellectual disability. Leiden (The Netherlands): International Association of Scientific Studies on Intellectual Disability, Special Interest Group on Health Issues; 1997.

Feightner JW. Screening for prostate cancer. In: Canadian Task Force on the Periodic Health Examination, editor(s). Canadian guide to clinical preventive health care. Ottawa (ON): Health Canada; 1994. p. 892-8.

Friedman RI. Use of advance directives: facilitating health care decisions by adults with mental retardation and their families. Ment Retard. 1998 Dec;36(6):444-56. PubMed

Fudge Schormans A, Sobsey D. Maltreatment of children with developmental disabilities. In: Brown I, Percy M, editor(s). A comprehensive guide to intellectual and developmental disabilities. Baltimore (MD): Paul H. Brookes Publishing Co.; 2007. p. 467-87.

Glassman P, Miller C. Dental disease prevention and people with special needs. J Calif Dent Assoc. 2003 Feb;31(2):149-60. PubMed

Hamilton S, Hankey CR, Miller S, Boyle S, Melville CA. A review of weight loss interventions for adults with intellectual disabilities. Obes

Rev. 2007 Jul;8(4):339-45. [61 references] PubMed

Haveman M, Heller T, Lee L, Maaskant M, Shooshtari S, Strydom A. Major health risks in aging persons with intellectual disabilities: an overview of recent studies. J Policy Pract Intellect Disabil. 2010;7(1):59-69.

Health Canada. Canadian guidelines for body weight classification in adults. [internet]. Ottawa (ON): Health Canada; 2003 [accessed 2009 Jul 14].

Healthy Living Unit, Public Health Agency of Canada. Canada's physical activity guide. [internet]. Ottawa (ON): Public Health Agency of Canada; 2003 [accessed 2009 Jul 14].

Henderson CM, Robinson LM, Davidson PW, Haveman M, Janicki MP, Albertini G. Overweight status, obesity, and risk factors for coronary heart disease in adults with intellectual disability. J Policy Pract Intellect Disabil. 2008;5(3):174-7.

Hollins S, Sinason V. Psychotherapy, learning disabilities and trauma: new perspectives. Br J Psychiatry. 2000 Jan;176:32-6. [32 references] PubMed

Hubert J, Hollins S. Men with severe learning disabilities and challenging behaviour in long-stay hospital care: qualitative study. Br J Psychiatry. 2006 Jan;188:70-4. PubMed

Hurley AD. Using the ABC sheet to analyze behavior: a training guide. Habilit Ment Healthc Newsl. 1997;16(5):81-9.

Iglar K, Katyal S, Matthew R, Dubey V. Complete health checkup for adults: Update on the Preventive Care Checklist Form. Can Fam Physician. 2008 Jan;54(1):84-8. [35 references] PubMed

Ismail AL, Lewis DW, Dingle JL. Prevention of periodontal disease. In: Canadian Task Force on the Periodic Health Examination, editor(s). Canadian guide to clinical preventive health care. Ottawa (ON): Health Canada; 1994. p. 420-31.

Kalachnik JE, Leventhal BL, James DH, Sovner R, Kastner TA, Walsk K, et al. Guidelines for the use of psychotropic medication. In: Reiss S, Aman MG, editor(s). Psychotropic medications and developmental disabilities: the international consensus handbook. Columbus (OH): Ohio State University, Nisonger Center; 1998. p. 45-72.

Kerr M, Guidelines Working Group, Scheepers M, Arvio M, Beavis J, Brandt C, Brown S, Huber B, Iivanainen M, Louisse AC, Martin P, Marson AG, Prasher V, Singh BK, Veendrick M, Wallace RA. Consensus guidelines into the management of epilepsy in adults with an intellectual disability. J Intellect Disabil Res. 2009 Aug;53(8):687-94. PubMed

Langley JM, Faughnan ME. Prevention of influenza in the general population: recommendation statement from the Canadian Task Force on Preventive Health Care. CMAJ. 2004 Nov 9;171(10):1169-70. [11 references] PubMed

Leddin D, Hunt R, Champion M, Cockeram A, Flook N, Gould M, Kim YI, Love J, Morgan D, Natsheh S, Sadowski D, Canadian Association of Gastroenterology, Canadian Digestive Health Foundation. Canadian Association of Gastroenterology and the Canadian Digestive Health Foundation: Guidelines on colon cancer screening. Can J Gastroenterol. 2004 Feb;18(2):93-9. PubMed

Lennox N, Bain C, Rey-Conde T, Purdie D, Bush R, Pandeya N. Effects of a comprehensive health assessment programme for Australian

adults with intellectual disability: a cluster randomized trial. Int J Epidemiol. 2007 Feb;36(1):139-46. PubMed

Lennox N, Developmental Disability Steering Group. Preventive health care and health promotion. In: Management guidelines: developmental disability. Version 2. Melbourne, Australia: Therapeutic Guidelines; 2005. p. 95-9.

Lewis DW, Ismail AI. Prevention of dental caries. In: Canadian Task Force on the Periodic Health Examination, editor(s). Canadian guide to clinical preventive health care. Ottawa (ON): Health Canada; 1994. p. 407-17.

Marks B, Sisirak J, Heller T. Health matters: the exercise, nutrition, and health education curriculum for people with developmental disabilities. [CD-ROM]. Baltimore (MD): Paul H. Brookes Publishing Co.; 2010

Mast EE, Weinbaum CM, Fiore AE, Alter MJ, Bell BP, Finelli L, Rodewald LE, Douglas JM Jr, Janssen RS, Ward JW, Advisory Committee on Immunization Practices (ACIP) Centers for Disease Control and Prevention. A comprehensive immunization strategy to eliminate transmission of hepatitis B virus infection in the United States: recommendations of the Advisory Committee on Immunization Practices (ACIP) Part II: immunization of adults. MMWR Recomm Rep. 2006 Dec 8;55(RR-16):1-33; quiz CE1-4. [3 references] PubMed

McCabe MP, McGillivray JA, Newton DC. Effectiveness of treatment programmes for depression among adults with mild/moderate intellectual disability. J Intellect Disabil Res. 2006 Apr;50(Pt 4):239-47. PubMed

McElduff A, Beange H. Men's health and well-being: testosterone deficiency. J Intellect Dev Disabil. 2003;28(2):211-3.

McElduff A, Center J, Beange H. Hypogonadism in men with intellectual disabilities: a population study. J Intellect Dev Disabil. 2003;28(2):163-70.

McGinnity M, Banks R, Barnes G, Frankish P, Hollins S, Hutchinson D, et al. Psychotherapy and learning disability. London (UK): Royal College of Psychiatrists; 2004.

Ministry of Community and Social Services. Community networks of specialized care. [internet]. Toronto (ON): Community Networks of Specialized Care; 2009 [accessed 2009 Jul 21].

Ministry of Community and Social Services. Our regional offices. Programs for adults with a developmental disability. [internet]. Toronto (ON): Ministry of Community and Social Services; 2008 [accessed 2009 Sep 17].

Moeschler JB. Genetic evaluation of intellectual disabilities. Semin Pediatr Neurol. 2008 Mar;15(1):2-9. PubMed

Mohr C, Costello H. Mental health assessment and monitoring tools for people with intellectual disabilities. In: Bouras N, Hold G, editor(s). Psychiatric and behavioural disorders in intellectual and developmental disabilities. 2nd ed. Cambridge (UK): Cambridge University Press; 2007. p. 24-41.

Morad M, Nelson NP, Merrick J, Davidson PW, Carmeli E. Prevalence and risk factors of constipation in adults with intellectual disability in residential care centers in Israel. Res Dev Disabil. 2007 Nov-Dec;28(6):580-6. PubMed

Morrison BJ. Screening for breast cancer. In: Canadian Task Force on the Periodic Health Examination, editor(s). Canadian guide to clinical preventive health care. Ottawa (ON): Health Canada; 1994. p. 788-95.

Moss S, Prosser H, Costello H, Simpson N, Patel P, Rowe S, Turner S, Hatton C. Reliability and validity of the PAS-ADD Checklist for detecting psychiatric disorders in adults with intellectual disability. J Intellect Disabil Res. 1998 Apr;42 (Pt 2):173-83. PubMed

National Advisory Committee on Immunization (NACI). Statement on human papillomavirus vaccine. An Advisory Committee Statement (ACS). Can Commun Dis Rep. 2007 Feb 15;33(ACS-2):1-31. PubMed

National Advisory Committee on Immunization. Canadian immunization guide. 7th ed. Ottawa (ON): Public Health Agency of Canada; 2006.

O'Brien G. Behavioural phenotypes in clinical practice. London (UK): Mac Keith; 2002.

O'Neil ME, Fragala-Pinkham MA, Westcott SL, Martin K, Chiarello LA, Valvano J, Rose RU. Physical therapy clinical management recommendations for children with cerebral palsy - spastic diplegia: achieving functional mobility outcomes. Pediatr Phys Ther. 2006 Spring;18(1):49-72. [289 references] PubMed

Perez-Achiaga N, Nelson S, Hassiotis A. Instruments for the detection of depressive symptoms in people with intellectual disabilities: a systematic review. J Intellect Disabil. 2009 Mar;13(1):55-76. [20 references] PubMed

Quint EH, Elkins TE. Cervical cytology in women with mental retardation. Obstet Gynecol. 1997 Jan;89(1):123-6. PubMed

Regnard C, Reynolds J, Watson B, Matthews D, Gibson L, Clarke C. Understanding distress in people with severe communication difficulties: developing and assessing the Disability Distress Assessment Tool (DisDAT). J Intellect Disabil Res. 2007 Apr;51(Pt 4):277-92. PubMed

Regnard D, Matthews D, Gibson L, Learning Disability and Palliative Care Team at Northgate Hospital. DisDAT: Disability Distress Assessment Tool. Northumberland (UK): Northumberland Tyne & Wear Trust, St. Oswald's Hospice; 2008.

Roland PS, Smith TL, Schwartz SR, Rosenfeld RM, Ballachanda B, Earll JM, Fayad J, Harlor AD Jr, Hirsch BE, Jones SS, Krouse HJ, Magit A, Nelson C, Stutz DR, Wetmore S. Clinical practice guideline: cerumen impaction. Otolaryngol Head Neck Surg. 2008 Sep;139(3 Suppl 2):S1-21. [97 references] PubMed

Royal College of Psychiatrists, British Psychological Society. Dementia and people with learning disabilities: guidance on the assessment, diagnosis, treatment and support of people with learning disabilities who develop dementia. London (UK): Royal College of Psychiatrists, British Psychological Society; 2009.

Rush AJ, Frances A. Guideline 2: informed consent. Expert consensus guideline series: treatment of psychiatric and behavioral problems in mental retardation. Am J Ment Retard. 2000;105(3):169.

Rush AJ, Frances A. Guideline 4: medication treatment: general principles. Expert consensus guideline series: treatment of psychiatric and behavioral problems in mental retardation. Am J Ment Retard. 2000;105(3):178-9.

Sequeira H, Howlin P, Hollins S. Psychological disturbance associated with sexual abuse in people with learning disabilities. Case-control study. Br J Psychiatry. 2003 Nov;183:451-6. PubMed

Society for the Study of Behavioural Phenotypes. SSBP syndrome information sheets. [internet]. Cambridge (UK): SSBP Online; [accessed

2009 Jul 18].

Sullivan SG, Hussain R, Threlfall T, Bittles AH. The incidence of cancer in people with intellectual disabilities. Cancer Causes Control. 2004 Dec;15(10):1021-5. PubMed

Summers J, Boyd K, Reid J, Adamson J, Habjan B, Gignac V, et al. The interdisciplinary mental health team. In: Griffiths DM, Stavrakaki C, Summers J, editor(s). Dual diagnosis; an introduction to the mental health needs of persons with developmental disabilities. Sudbury (ON): Habilitative Mental Health Resource Network; 2002. p. 325-57.

Summers J, Stavrakaki C, Griffiths D, Cheetham T. Comprehensive screening and assessment. In: Griffiths DM, Stavrakaki C, Summers J, editor(s). Dual diagnosis; an introduction to the mental health needs of persons with developmental disabilities. Sudbury (ON): Habilitative Mental Health Resource Network; 2002. p. 151-92.

Taylor JL, Lindsay WR, Willner P. CBT for people with intellectual disabilities: emerging evidence, cognitive ability and IQ effects. Behav Cogn Psychother. 2008;36(6):723-33.

The AAIDD Ad Hoc Committee on Terminology and Classification. Adaptive behavior and its assessment. In: Intellectual disability: definition, classification, and systems of supports. 11th ed. Washington (DC): American Association on Intellectual and Developmental Disabilities; 2010. p. 43-55.

Tuffrey-Wijne I, McEnhill L. Communication difficulties and intellectual disability in end-of-life care. Int J Palliat Nurs. 2008 Apr;14(4):189-94. [23 references] PubMed

Tyrer P, Oliver-Africano PC, Ahmed Z, Bouras N, Cooray S, Deb S, Murphy D, Hare M, Meade M, Reece B, Kramo K, Bhaumik S, Harley D, Regan A, Thomas D, Rao B, North B, Eliahoo J, Karatela S, Soni A, Crawford M. Risperidone, haloperidol, and placebo in the treatment of aggressive challenging behaviour in patients with intellectual disability: a randomised controlled trial. Lancet. 2008 Jan 5;371(9606):57-63. PubMed

University of Washington. GeneReviews: medical genetics information resouce. [database online]. Seattle (WA): University of Washington; 1993 [accessed 2009 Apr 14].

van Schrojenstein Lantman-de Valk HM, Walsh PN. Managing health problems in people with intellectual disabilities. BMJ. 2008;337:a2507. [24 references] PubMed

Wallace R, Schluter Pj, Duff M, Ouellette-Kuntz H, Webb PM. A review of the risk factors for, consequences, diagnosis, and management of Helicobacter pylori in adults with intellectual disabilities. J Policy Pract Intellect Disabil. 2004;1(3-4):147-63.

Wallace RA, Schluter P. Audit of cardiovascular disease risk factors among supported adults with intellectual disability attending an ageing clinic. J Intellect Dev Disabil. 2008 Mar;33(1):48-58. PubMed

Wilkinson JE, Cerreto MC. Primary care for women with intellectual disabilities. J Am Board Fam Med. 2008 May-Jun;21(3):215-22. [59 references] PubMed

Wilson W, Taubert KA, Gewitz M, Lockhart PB, Baddour LM, Levison M, Bolger A, Cabell CH, Takahashi M, Baltimore RS, Newburger

JW, Strom BL, Tani LY, Gerber M, Bonow RO, Pallasch T, Shulman ST, Rowley AH, Burns JC, Ferrieri P, Gardner T, Goff D, Durack DT, American Heart Association Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, Quality of Care and Outcomes Research Interdisciplinary Working Group, American Dental Association. Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis and Kawasaki Disease Committee [trunc]. J Am Dent Assoc. 2007 Jun;138(6):739-45, 747-60. [147 references] PubMed

Zylstra RG, Porter LL, Shapiro JL, Prater CD. Prevalence of osteoporosis in community-dwelling individuals with intellectual and/or developmental disabilities. J Am Med Dir Assoc. 2008 Feb;9(2):109-13. PubMed

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate primary care of adults with developmental disabilities (DD), which may lead to:

- Improved quality of primary care of adults with DD
- Improved access to health care
- Improved state of health
- Prevention of suffering, morbidity, and premature death

Potential Harms

- Psychotropic medications can be problematic for adults with developmental disabilities (DD) and should therefore be used judiciously. Patients might be taking multiple medications and can thus be at increased risk of adverse medication interactions. Some adults with DD might have atypical responses or side effects at low doses.
- Antipsychotic medications increase risk of metabolic syndrome and can have other serious side effects (e.g., akathisia, cardiac conduction problems, swallowing difficulties, bowel dysfunction).

Qualifying Statements

Qualifying Statements

Limitations

- The aim of the original guideline document is to inform primary care providers of the most prevalent health issues of adults with developmental disabilities (DD) as a group and of the best approaches to management. However, any such set of guidelines will always be limited in their application by the reality that adults with DD are not a homogeneous group nor do they experience health disorders in the same way. These guidelines are not meant to replace attentive observation and prudent clinical decisions. The most appropriate care for an adult with DD takes into account relevant factors in his or her particular circumstances. Furthermore, although these guidelines are generally applicable to adults with DD, primary care providers will need to address additional specific health issues when there is a known cause of DD.
- The division of these guidelines into distinct physical, behavioural, and mental health categories was intended to facilitate their application. Several guidelines, however, address the interaction of physical factors with behavioural and mental health ones, and between these and environmental factors and other determinants of health.

- Some of these updated guidelines recommend the use of resources and specialized services that, while generally available in Canada, might be lacking or inaccessible in some regional health service systems. In such circumstances, it is necessary to adapt these guidelines to allow primary care providers to provide a reasonable standard of care and to develop practical resource-sharing strategies (e.g., using clinical videoconferencing).

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

Staying Healthy

IOM Domain

Effectiveness

Patient-centeredness

Safety

Identifying Information and Availability

Bibliographic Source(s)

Sullivan WF, Berg JM, Bradley E, Cheetham T, Denton R, Heng J, Hennen B, Joyce D, Kelly M, Korossy M, Lunsky Y, McMillan S. Primary care of adults with developmental disabilities: Canadian consensus guidelines. Can Fam Physician. 2011 May;57(5):541-53, e154-68. [185 references] PubMed

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2011 May

Guideline Developer(s)

College of Family Physicians of Canada - Professional Association

Source(s) of Funding

College of Family Physicians of Canada

Guideline Committee

Not stated

Composition of Group That Authored the Guideline

Authors: William F. Sullivan, MD, CCFP, PhD, family physician, St Michael's Hospital and Surrey Place Centre in Toronto, Ont, Associate Professor, University of Toronto, bioethicist; Joseph M. Berg, MB, BCh, MSc, FRCPsych, FCCMG, Professor Emeritus, Faculty of Medicine, University of Toronto, medical consultant, Surrey Place Centre; Elspeth Bradley, PhD, MB BS, FRCPC, FRCPsych, intellectual disabilities psychiatrist, Associate Professor, Department of Psychiatry, University of Toronto, Psychiatrist-in-Chief, Surrey Place Centre; Tom Cheetham, MD, CCFP, family physician, Surrey Place Centre; Richard Denton, MD, CCFP, FCFP, FRRMS, family physician, Kirkland Lake, Ont, Assistant Professor, Clinical Science, Department of Family Medicine, Northern Ontario School of Medicine; John Heng, MA, bioethicist in philosophy and thanatology, King's University College, University of Western Ontario, London, Ont; Brian Hennen, MA, MD, CCFP, Professor of Family Medicine, Dalhousie University, Halifax, NS; David Joyce, MD, CCFP, family physician, clinical investigator, Department of Family Medicine, University of British Columbia, Vancouver, BC; Maureen Kelly, RN, MPA, Project Coordinator, Developmental Disabilities Primary Care Initiative, Surrey Place Centre; Marika Korossy, librarian, Surrey Place Centre; Yona Lunsky, PhD, CPsych, Clinician Scientist, Centre for Addiction and Mental Health, Toronto, Adjunct Scientist, Institute for Clinical and Evaluative Sciences, Associate Professor, University of Toronto; Shirley McMillan, RN, MN, CDDN, Mental Health Nurse Specialist, Surrey Place Centre

Financial Disclosures/Conflicts of Interest

None declared

Guideline Status

This is the current release of the guideline.

Guideline Availability

Electronic copies: Available from the Canadian Family Physician Journal Web site .

Availability of Companion Documents

None available

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI Institute on June 27, 2013. This summary was updated by ECRI Institute on May 24, 2016 following the U.S. Food and Drug Administration advisory on Olanzapine.

Copyright Statement

Disclaimer

NGC Disclaimer